Endovascular repair of multiple infrageniculate aneurysms in a patient with vascular type Ehlers-Danlos syndrome

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Patients with vascular type Ehler-Danlos syndrome can develop aneurysms in unusual locations. We describe the case of a 33-year-old woman with vascular type Ehlers-Danlos syndrome who developed metachronous tibial artery aneurysms that were sequentially treated with endovascular means. (J Vasc Surg 2011;54:848-50.)

Patients with vascular type Ehlers-Danlos syndrome (EDS), previously Ehlers-Danlos type IV, can develop aneurysms in unusual locations.[1] Unfortunately, vascular rupture or dissection is a common presentation reported to occur in 77% of affected patients.[1,2] We describe the case of a young woman with vascular EDS who developed metachronous tibial artery aneurysms sequentially treated with coil embolization and stent graft exclusion.

CASE REPORT

A 33-year-old woman first presented to our institution in 2001 with severe abdominal pain following a recent appendectomy at an outside hospital. Her past medical history was significant for coronary artery disease of unclear etiology, myocardial infarction, and factor V Leiden deficiency. She had no known family history of collagen vascular disease. An exploratory laparotomy was performed under the presumptive diagnosis of intestinal ischemia. The laparotomy was negative, but 6 days into recovery, the patient was re-explored for shock and abdominal distention. A ruptured splenic artery aneurysm was noted and treated with ligation and splenectomy.

Within 1 week, the patient developed acute swelling of her left arm with a compartment syndrome secondary to rupture of an aneurysm of a large branch of the brachial artery. This was temporally related to a blood pressure cuff inflation and was treated with ligation and compartment release. A skin biopsy was performed for suspected vasculitis, and biochemical testing of fibroblast collagen secretion, followed by DNA evaluation, confirmed the diagnosis of vascular EDS.

Eight months later, the patient was diagnosed with bilateral anterior tibial artery aneurysms, which were successfully coil embolized in a staged fashion. Her posterior tibial and peroneal vessels were not aneurismal at that time.

Seven years after her initial presentation, the patient reported to the emergency department with hypotension and a spontaneous, painful, rapidly expanding lump in her right upper thigh. Emergency surgery revealed a hemorrhaging venous malformation at and beyond the level of the femoral vein confluence with the saphenous vein. Pressure control was obtained and wide suture ligation performed. Postoperatively, the patient developed deep vein thrombosis of the right lower extremity and was anticoagulated with coumadin. Her lower extremity swelling resolved with compression stockings.

The patient was also known to have a left posterior tibial artery (PTA) aneurysm (Fig 1). Over 2 years, duplex ultrasound evaluation demonstrated an increase in diameter from 2 to 3 cm at which time the patient developed a pulsatile calf mass on clinical examination. Though asymptomatic, need for repair was based on the absolute size and enlarging diameter. To maintain two-vessel runoff to the foot, the decision was made to attempt stent graft repair, given that the diameter of the nonaneurismal portion of the PTA was large enough and was 4 mm by CT measurement. Through contralateral up and over transfemoral access with a 7F sheath, a 5 × 10 mm self-expanding Viabahn stent graft (W. L. Gore and Associates, Flagstaff, Ariz) was deployed across the aneurysm into the PTA outflow. Poststenting angioplasty with a 5 mm balloon was performed. A completion angiogram showed good apposition of the stent and complete exclusion of the aneurysm (Fig 2). Access site hemostasis was achieved with manual compression after immediate failure of attempted closure with an 8F angioseal (Saint Jude Medical, St. Paul, Minn). The patient developed a hematoma at the puncture site that spontaneously resolved and was acutely investigated by duplex imaging and CT scan. She maintained a palpable PTA pulse through her hospital stay and was discharged home on clopidogrel. No evidence of pseudoaneurysm was noted on follow-up duplex scanning at 1 and 4 weeks. At 1-month follow-up, the PTA pulse was lost, and the stent graft was completely occluded by duplex ultrasound, with a patent PTA at the ankle but no flow into the PTA aneurysm. The patient was asymptomatic with normal ambulation status. At 1-year follow-up, the patient continued to be asymptomatic, with an excluded PTA aneurysm.

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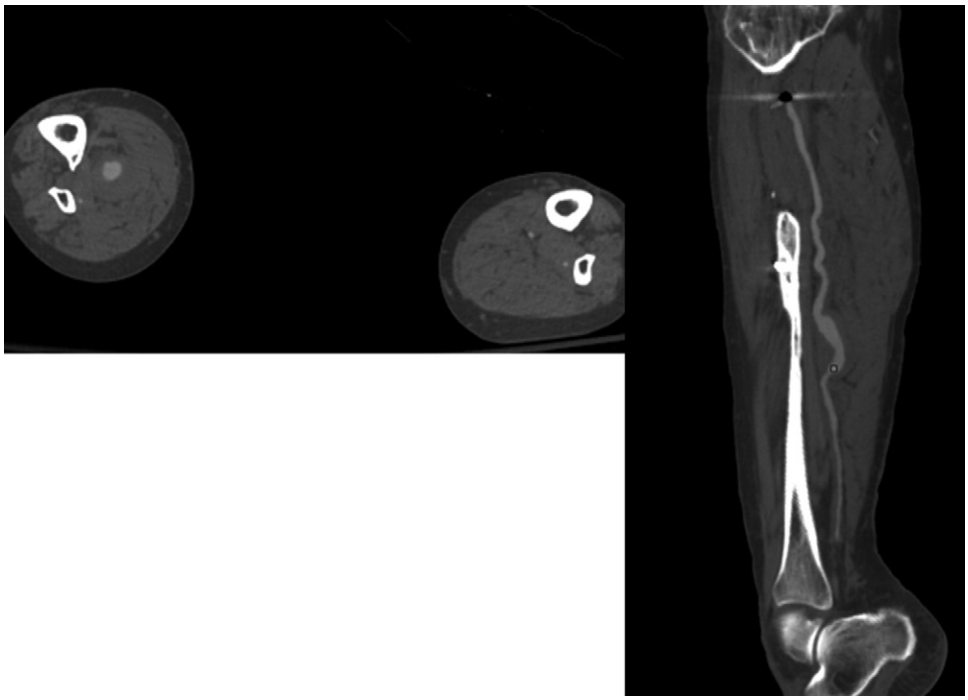

Fig 1. Computed tomography angiography (CTA) of left lower extremity demonstrating a 3 cm posterior tibial artery (PTA) aneurysm.

DISCUSSION

Infrapopliteal aneurysms are exceedingly rare and have a poorly characterized natural history.[3] Prior to this report, only 41 true aneurysms below the popliteal fossa have been reported in the English literature,[3-10] 14 of those involving posterior tibial arteries,[5] and none with concomitant vascular EDS. Since rupture is rare,[5] small and asymptomatic infrapopliteal aneurysms may be observed safely, while symptomatic aneurysms require prompt surgical intervention, most commonly direct repair or ligation.[5,6,11]

These generalizations must be carefully evaluated in light of the overall vascular fragility of patients with vascular EDS.[2] In fact, 44% of vascular EDS patients with arterial rupture die before repair, necessitating strategic management of vascular complications prior to rupture.[12] In light of our patient's history of multiple ruptured aneurysms prior to development of her PTA aneurysm, and given the size and enlarging diameter of her aneurysm, intervention was decided.

Since a single vessel runoff is sufficient if the pedal arch is patent, it is generally acceptable to sacrifice an aneurismal tibial vessel.[10] As in severe atherosclerotic disease associated with tibial aneurysm,[11] this practice also requires personalized deliberation in a patient with vascular EDS. Our patient had already lost ATA runoff from prior coil embolization, and coiling of her PTA aneurysm would leave her with a single vessel runoff that is potentially at risk for future aneurismal degeneration. While stent graft repair is generally accepted for popliteal aneurysms,[13] it is not well established for tibial aneurysms. Only four accounts of stent graft repair of an ATA pseudoaneurysm and one of a PTA pseudoaneurysm, all successful, have been reported in the English literature.[14] Stents used included JOSTENT (Abbott Vascular Devices, Abbot Park, Ill) or Symbiot stent graft (Boston Scientific, Natick, Mass),[14] and only short-term patency was reported.

No reports exist describing stent grafting of true tibial artery aneurysms or of vascular EDS complications.[2] Moreover, stenting presents a potential risk of arterial wall injury and pseudoaneurysm development at the access site in these patients.[14] Nevertheless, endovascular repair was chosen due to the potential advantage of maintaining a two-vessel runoff and given the possibility of future peroneal aneurysm formation. The Viabahn (W. L. Gore and Associates) stent was selected because of the suitable large landing zone diameter, the stent's proven performance in popliteal aneurysms, and its superior flexibility compared with balloon expandable stent grafts. Surgical bypass was not considered given the associated risks in vascular EDS patients in general[1] and the patient's history of symptomatic coronary artery disease.

Although tibial artery stent grafting has been described, the durability of covered stents in this vascular bed is not well characterized and could be adversely affected by small target vessel diameter and excessive mobility from the calf musculature. This could explain the graft failure in this patient, along with excessive vessel tortuosity and her coexisting hypercoagulable state. While coiling is probably the safest intervention in vascular EDS patients,[2] stent

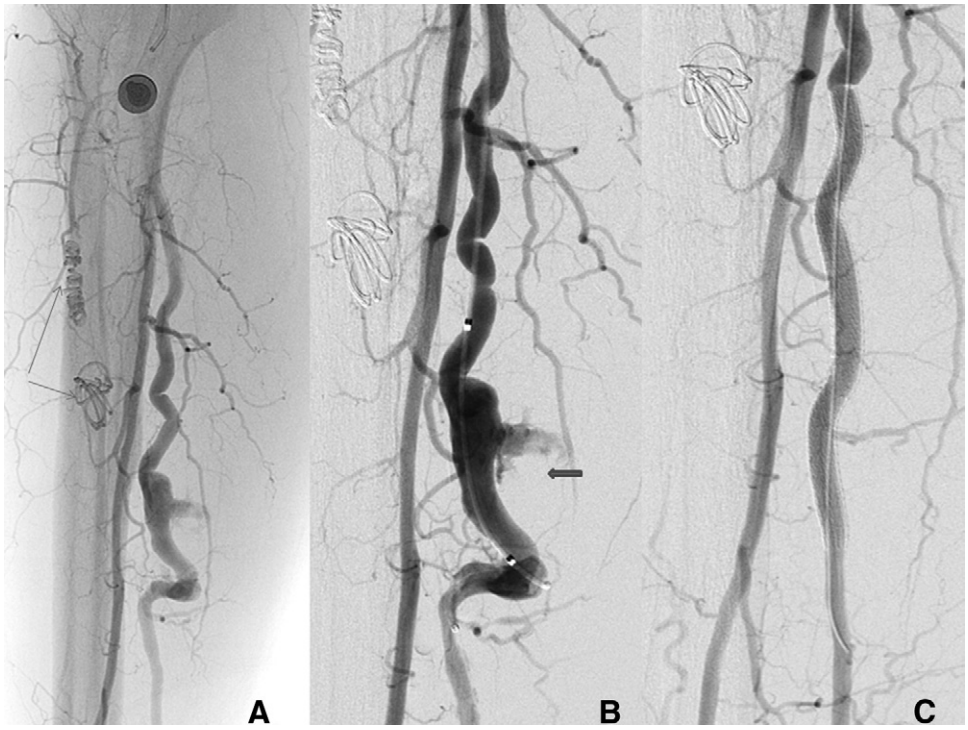

Fig 2. A, Initial angiogram demonstrating a posterior tibial artery (PTA) aneurysm with coils noted (*small arrows*) in the anterior tibial artery distribution at the site of a previous aneurysm. **B,** A large PTA aneurysm is noted with mural thrombus (*large arrow*) and significant outflow tortuosity. **C,** Complete exclusion of the PTA aneurysm is noted following deployment of a Viabahn (W. L. Gore and Associates) stent graft.

grafting is a consideration when faced with preservation of runoff and limb viability. Given her history, this patient has the potential to develop future tibial aneurysms requiring intervention, making maintenance of a two-vessel runoff a sensible goal.

CONCLUSION

Patients with vascular EDS can present with multiple tibial artery aneurysms, limiting the options of runoff preservation. Endovascular treatment is safe and can be achieved with coil embolization. Stent grafting may be attractive to maximize runoff, but patency of current grafts in the tibial bed is unproven.

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